



Complete Summary

GUIDELINE TITLE

Tetralogy of Fallot. In: ACC/AHA 2008 guidelines for the management of adults with congenital heart disease. A report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (Writing Committee to Develop Guidelines on the Management of Adults With Congenital Heart Disease).

BIBLIOGRAPHIC SOURCE(S)

Warnes CA, Williams RG, Bashore TM, Child JS, Connolly HM, Dearani JA, del Nido P, Fasules JW, Graham TP, Hijazi ZM, Hunt SA, King ME, Landzberg MJ, Miner PD, Radford MJ, Walsh EP, Webb GD, Smith SC Jr, Jacobs AK, Adams CD, Anderson JL, Antman EM, Buller CD, Creager MA, Ettinger SM, Halperin JL, Hunt SA, Krumholz HM, Kushner FG, Lytle BW, Nishimura RA, Page RL, Riegel B, Tarkington LG, Yancy CW. Tetralogy of Fallot. In: ACC/AHA 2008 guidelines for the management of adults with congenital heart disease. J Am Coll Cardiol 2008;52(23):e215-21.

GUIDELINE STATUS

This is the current release of the guideline.

The guidelines will be reviewed annually by the American College of Cardiology/American Heart Association (ACC/AHA) Task Force on Practice Guidelines and considered current unless they are updated, revised, or withdrawn from distribution.

COMPLETE SUMMARY CONTENT

SCOPE
METHODOLOGY - including Rating Scheme and Cost Analysis
RECOMMENDATIONS
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IDENTIFYING INFORMATION AND AVAILABILITY

SCOPE

DISEASE/CONDITION(S)

- Adult congenital heart disease
- Tetralogy of Fallot

GUIDELINE CATEGORY

Counseling
Diagnosis
Evaluation
Management
Risk Assessment
Screening
Treatment

CLINICAL SPECIALTY

Cardiology
Family Practice
Internal Medicine
Radiology
Thoracic Surgery

INTENDED USERS

Health Care Providers
Physicians

GUIDELINE OBJECTIVE(S)

- To assist healthcare providers in clinical decision making by describing a range of generally acceptable approaches for diagnosis, management, and prevention of specific diseases or conditions associated with adult congenital heart disease (ACHD)
- To define practices that meet the needs of most patients in most circumstances
- To support the practicing cardiologist in the care of ACHD patients by providing a consensus document that outlines the most important diagnostic and management strategies and indicates when referral to a highly specialized center is appropriate

TARGET POPULATION

Adults with congenital heart disease and tetralogy of Fallot

INTERVENTIONS AND PRACTICES CONSIDERED

Diagnosis/Evaluation

1. Echocardiographic examinations
2. Magnetic resonance Imaging (MRIs)
3. Screening for heritable causes of tetralogy of Fallot
4. Cardiac catheterization

5. Assessment of coronary artery anatomy

Management/Treatment

1. Pulmonary valve replacement
2. Interventional catheterization
3. Preoperative or intraoperative stenting or intraoperative patch angioplasty

Follow-Up for Arrhythmias

1. Annual surveillance
2. History
3. Electrocardiogram
4. Assessment of right ventricular (RV) function
5. Periodic exercise testing
6. Periodic Holter monitoring

MAJOR OUTCOMES CONSIDERED

- Late survival after tetralogy of Fallot repair
- Incidence of sudden cardiac death after tetralogy of Fallot surgery

METHODOLOGY

METHODS USED TO COLLECT/SELECT EVIDENCE

Hand-searches of Published Literature (Primary Sources)
Hand-searches of Published Literature (Secondary Sources)
Searches of Electronic Databases

DESCRIPTION OF METHODS USED TO COLLECT/SELECT THE EVIDENCE

Unlike other American College of Cardiology/American Heart Association (ACC/AHA) practice guidelines; there is not a large body of peer-reviewed published evidence to support most recommendations, which will be clearly indicated in the text. An extensive literature survey was conducted that led to the incorporation of 647 references. Searches were limited to studies, reviews, and other evidence conducted in human subjects and published in English. Key search words included but were not limited to adult congenital heart disease (ACHD), atrial septal defect, arterial switch operation, bradycardia, cardiac catheterization, cardiac reoperation, coarctation, coronary artery abnormalities, cyanotic congenital heart disease, Doppler-echocardiography, d-transposition of the great arteries, Ebstein's anomaly, Eisenmenger physiology, familial, heart defect, medical therapy, patent ductus arteriosus, physical activity, pregnancy, psychosocial, pulmonary arterial hypertension, right heart obstruction, supraaortic pulmonary stenosis, surgical therapy, tachyarrhythmia, tachycardia, tetralogy of Fallot, transplantation, tricuspid atresia, and Wolff-Parkinson-White. Additionally, the writing committee reviewed documents related to the subject matter previously published by the ACC and AHA. References selected and published in this document are representative and not all-inclusive.

NUMBER OF SOURCE DOCUMENTS

Not stated

METHODS USED TO ASSESS THE QUALITY AND STRENGTH OF THE EVIDENCE

Weighting According to a Rating Scheme (Scheme Given)

RATING SCHEME FOR THE STRENGTH OF THE EVIDENCE

Applying Classification of Recommendations and Level of Evidence

		SIZE OF TREATMENT EFFECT		
		CLASS I <i>Benefit >>> Risk</i> Procedure/Treatment SHOULD be performed/administered	CLASS IIa <i>Benefit >> Risk</i> <i>Additional studies with focused objectives needed</i> IT IS REASONABLE to perform procedure/administer treatment	CLASS IIb <i>Benefit ≥ Risk</i> <i>Additional studies with focused objectives needed</i> <i>registry data would be helpful</i> Procedure/Treatment MAY BE CONSIDERED
Estimate of Certainty (Precision) of Treatment Effect	LEVEL A Multiple population evaluated* Data derived from multiple randomized clinical trials or meta-analyses	<ul style="list-style-type: none"> Recommendation that procedure or treatment is useful/effective Sufficient evidence from multiple randomized trials or meta-analyses 	<ul style="list-style-type: none"> Recommendation in favor of treatment of procedure being useful/effective Some conflicting evidence from multiple randomized trials or meta-analyses 	<ul style="list-style-type: none"> Recommendation in favor of treatment of procedure being useful/effective Greater evidence from multiple randomized trials or meta-analyses
	LEVEL B Limited population evaluated* Data derived from a single randomized clinical trial or nonrandomized studies	<ul style="list-style-type: none"> Recommendation that procedure or treatment is useful/effective Evidence from single randomized trial or nonrandomized studies 	<ul style="list-style-type: none"> Recommendation in favor of treatment of procedure being useful/effective Some conflicting evidence from single randomized trial or nonrandomized studies 	<ul style="list-style-type: none"> Recommendation in favor of treatment of procedure being useful/effective Greater evidence from single randomized trial or nonrandomized studies
	LEVEL C	<ul style="list-style-type: none"> Recommendation 	<ul style="list-style-type: none"> Recommendation in 	<ul style="list-style-type: none"> Recommendation

		SIZE OF TREATMENT EFFECT		
	Very limited population evaluated*	<ul style="list-style-type: none"> that procedure or treatment is useful/effective Only expert opinion, case studies, or standard-of-care 	<ul style="list-style-type: none"> favor of treatment of procedure being useful/effective Only diverging expert opinion, case studies, or standard-of-care 	<ul style="list-style-type: none"> useful or less well supported Only diverging expert opinion or standard-of-care
	Only consensus opinion of experts, case studies or standard of care.			

*Data available from clinical trials or registries about the usefulness/efficacy in different subpopulations, such as gender, age, history of diabetes, history of prior myocardial infarction, history of heart failure, and prior aspirin use. A recommendation with Level of Evidence B or C does not imply that the recommendation is weak. Many important clinical questions addressed in the guidelines do not lend themselves to clinical trials. Even though randomized trials are not available, there may be a very clear clinical consensus that a particular test or therapy is useful or effective.

Note: In 2003, the American College of Cardiology/American Heart Association (ACC/AHA) Task Force on Practice Guidelines developed a list of suggested phrases to use when writing recommendations. All guideline recommendations have been written in full sentences that express a complete thought, such that a recommendation, even if separated and presented apart from the rest of the document (including headings above sets of recommendations), would still convey the full intent of the recommendation. It is hoped that this will increase readers' comprehension of the guidelines and will allow queries at the individual recommendation level. (See Table 1 in the original guideline document for a list of suggested phrases for writing recommendations.)

METHODS USED TO ANALYZE THE EVIDENCE

Systematic Review

DESCRIPTION OF THE METHODS USED TO ANALYZE THE EVIDENCE

The committee reviewed and ranked evidence supporting current recommendations with the weight of evidence ranked as Level A if the data were derived from multiple randomized clinical trials involving a large number of individuals. The committee ranked available evidence as Level B when data were derived from a limited number of trials involving a comparatively small number of patients or from well-designed data analyses of nonrandomized studies or observational data registries. Evidence was ranked as Level C when the consensus of experts was the primary source of the recommendation. In the narrative portions of these guidelines, evidence is generally presented in chronological order of development. Studies are identified as observational, randomized, prospective, or retrospective. The committee emphasizes that for certain conditions for which no other therapy is available, the indications are based on expert consensus and years of clinical experience and are thus well supported, even though the evidence was ranked as Level C. An analogous example is the use of penicillin in pneumococcal pneumonia where there are no randomized trials and only clinical experience. When indications at Level C are supported by historical clinical data, appropriate references (e.g., case reports and clinical reviews) are cited if

available. When Level C indications are based strictly on committee consensus, no references are cited. The final recommendations for indications for a diagnostic procedure, a particular therapy, or an intervention in adult congenital heart disease (ACHD) patients summarize both clinical evidence and expert opinion. The schema for classification of recommendations and level of evidence illustrates how the grading system provides an estimate of the size of treatment effect and an estimate of the certainty of the treatment effect (see "Rating Scheme for the Strength of the Evidence" above).

METHODS USED TO FORMULATE THE RECOMMENDATIONS

Expert Consensus

DESCRIPTION OF METHODS USED TO FORMULATE THE RECOMMENDATIONS

The American College of Cardiology/American Heart Association (ACC/AHA) Task Force on Practice Guidelines was formed to create clinical practice guidelines for select cardiovascular conditions with important implications for public health. This guideline writing committee was assembled to adjudicate the evidence and construct recommendations regarding the diagnosis and treatment of adult congenital heart disease (ACHD). Writing committee members were selected with attention to ACHD subspecialties, broad geographic representation, and involvement in academic medicine and clinical practice. The writing committee included representatives of the American Society of Echocardiography, Heart Rhythm Society, International Society for Adult Congenital Heart Disease, Society for Cardiovascular Angiography and Interventions, and Society of Thoracic Surgeons.

Writing committees are specifically charged to perform a formal literature review, weigh the strength of evidence for or against particular treatments or procedures, and include estimates of expected health outcomes where data exist. Patient-specific modifiers, comorbidities, and issues of patient preference that might influence the choice of tests or therapies are considered, as well as the frequency of follow-up and cost-effectiveness. When available, information from studies on cost is considered, but data on efficacy and clinical outcomes constitute the primary basis for recommendations in these guidelines.

RATING SCHEME FOR THE STRENGTH OF THE RECOMMENDATIONS

See "Rating Scheme for the Strength of the Evidence" field, above.

COST ANALYSIS

A formal cost analysis was not performed and published cost analyses were not reviewed.

METHOD OF GUIDELINE VALIDATION

External Peer Review
Internal Peer Review

DESCRIPTION OF METHOD OF GUIDELINE VALIDATION

This document was reviewed by 3 external reviewers nominated from both the American College of Cardiology (ACC) and the American Heart Association (AHA), as well as reviewers from the American Society of Echocardiography, Canadian Cardiovascular Society, Heart Rhythm Society, International Society for Adult Congenital Heart Disease, and Society of Thoracic Surgeons, and 20 individual content reviewers which included reviewers from the ACC Congenital Heart Disease and Pediatric Cardiology Committee and the AHA Congenital Cardiac Defects Committee. All reviewer relationships with industry information were collected and distributed to the writing committee and are published in the original guideline document (see the "Conflicts of Interest/Financial Disclosures" field in this document).

This document was approved for publication by the governing bodies of the American College of Cardiology Foundation (ACCF) and the AHA and endorsed by the American Society of Echocardiography, Heart Rhythm Society, International Society for Adult Congenital Heart Disease, Society for Cardiovascular Angiography and Interventions, and Society of Thoracic Surgeons.

RECOMMENDATIONS

MAJOR RECOMMENDATIONS

The American College of Cardiology/American Heart Association (ACC/AHA) classification of the recommendations for patient evaluation and treatment (classes I-III) and the levels of evidence (A-C) are defined at the end of the "Major Recommendations" field.

Recommendations for Evaluation and Follow-Up of the Repaired Patient

Class I

1. Patients with repaired tetralogy of Fallot should have at least annual follow-up with a cardiologist who has expertise in adult congenital heart disease (ACHD). (**Level of Evidence: C**)
2. Patients with tetralogy of Fallot should have echocardiographic examinations and/or magnetic resonance imagings (MRIs) performed by staff with expertise in ACHD. (**Level of Evidence: C**)
3. Screening for heritable causes of their condition (e.g., 22q11 deletion) should be offered to all patients with tetralogy of Fallot. (**Level of Evidence: C**)
4. Before pregnancy or if a genetic syndrome is identified, consultation with a geneticist should be arranged for patients with tetralogy of Fallot. (**Level of Evidence: B**)
5. Patients with unrepaired or palliated forms of tetralogy should have a formal evaluation at an ACHD center regarding suitability for repair. (**Level of Evidence: B**)

Recommendations for Imaging

Class 1

1. Comprehensive echocardiographic imaging should be performed in a regional ACHD center to evaluate the anatomy and hemodynamics in patients with repaired tetralogy of Fallot. (**Level of Evidence: B**)

Recommendations for Diagnostic and Interventional Catheterization for Adults With Tetralogy of Fallot

Class I

1. Catheterization of adults with tetralogy of Fallot should be performed in regional centers with expertise in ACHD. (**Level of Evidence: C**)
2. Coronary artery delineation should be performed before any intervention for the right ventricular outflow tract (RVOT). (**Level of Evidence: C**)

Class IIb

1. In adults with repaired tetralogy of Fallot, catheterization may be considered to better define potentially treatable causes of otherwise unexplained left ventricle (LV) or right ventricle (RV) dysfunction, fluid retention, chest pain, or cyanosis. In these circumstances, transcatheter interventions may include:
 - a. Elimination of residual shunts or aortopulmonary collateral vessels. (**Level of Evidence: C**)
 - b. Dilation (with or without stent implantation) of RVOT obstruction. (**Level of Evidence: B**)
 - c. Elimination of additional muscular or patch margin ventricular septal defect (VSD). (**Level of Evidence: C**)
 - d. Elimination of residual atrial septal defect (ASD). (**Level of Evidence: B**)

Recommendations for Surgery for Adults With Previous Repair of Tetralogy of Fallot

Class I

1. Surgeons with training and expertise in congenital heart disease (CHD) should perform operations in adults with previous repair of tetralogy of Fallot. (**Level of Evidence: C**)
2. Pulmonary valve replacement is indicated for severe pulmonary regurgitation and symptoms or decreased exercise tolerance. (**Level of Evidence: B**)
3. Coronary artery anatomy, specifically the possibility of an anomalous anterior descending coronary artery across the RVOT, should be ascertained before operative intervention. (**Level of Evidence: C**)

Class IIa

1. Pulmonary valve replacement is reasonable in adults with previous tetralogy of Fallot, severe pulmonary regurgitation, and any of the following:
 - a. Moderate to severe right ventricular (RV) dysfunction. (**Level of Evidence: B**)
 - b. Moderate to severe RV enlargement. (**Level of Evidence: B**)

- c. Development of symptomatic or sustained atrial and/or ventricular arrhythmias. (**Level of Evidence: C**)
 - d. Moderate to severe tricuspid regurgitation (TR). (**Level of Evidence: C**)
- 2. Collaboration between ACHD surgeons and ACHD interventional cardiologists, which may include preoperative stenting, intraoperative stenting, or intraoperative patch angioplasty, is reasonable to determine the most feasible treatment for pulmonary artery stenosis. (**Level of Evidence: C**)
- 3. Surgery is reasonable in adults with prior repair of tetralogy of Fallot and residual RVOT obstruction (valvular or subvalvular) and any of the following indications:
 - a. Residual RVOT obstruction (valvular or subvalvular) with peak instantaneous echocardiography gradient greater than 50 mm Hg. (**Level of Evidence: C**)
 - b. Residual RVOT obstruction (valvular or subvalvular) with RV/LV pressure ratio greater than 0.7. (**Level of Evidence: C**)
 - c. Residual RVOT obstruction (valvular or subvalvular) with progressive and/or severe dilatation of the right ventricle with dysfunction. (**Level of Evidence: C**)
 - d. Residual ventricular septal defect (VSD) with a left-to-right shunt greater than 1.5:1. (**Level of Evidence: B**)
 - e. Severe aortic regurgitation (AR) with associated symptoms or more than mild LV dysfunction. (**Level of Evidence: C**)
 - f. A combination of multiple residual lesions (e.g., VSD and RVOT obstruction) leading to RV enlargement or reduced RV function. (**Level of Evidence: C**)

Recommendations for Interventional Catheterization

Class I

- 1. Interventional catheterization in an ACHD center is indicated for patients with previously repaired tetralogy of Fallot with the following indications:
 - a. To eliminate residual native or palliative systemic–pulmonary artery shunts. (**Level of Evidence: B**)
 - b. To manage coronary artery disease. (**Level of Evidence: B**)

Class IIa

- 1. Interventional catheterization in an ACHD center is reasonable in patients with repaired tetralogy of Fallot to eliminate a residual ASD or VSD with a left-to-right shunt greater than 1.5:1 if it is in an appropriate anatomic location. (**Level of Evidence: C**)

Key Issues to Evaluate and Follow-Up

Recommendations for Arrhythmias: Pacemaker/Electrophysiology Testing

Class I

1. Annual surveillance with history, electrocardiogram (ECG), assessment of RV function, and periodic exercise testing is recommended for patients with pacemakers/automatic implantable cardioverter defibrillators. (**Level of Evidence: C**)

Class IIa

1. Periodic Holter monitoring can be beneficial as part of routine follow-up. The frequency should be individualized depending on the hemodynamics and clinical suspicion of arrhythmia. (**Level of Evidence: C**)

Class IIb

1. Electrophysiology testing in an ACHD center may be reasonable to define suspected arrhythmias in adults with tetralogy of Fallot. (**Level of Evidence: C**)

Definitions:

Applying Classification of Recommendations and Level of Evidence

Â		SIZE OF TREATMENT EFFECT		
Â		CLASS I	CLASS IIa	CLASS IIb
		<i>Benefit >>> Risk</i> Procedure/Treatment SHOULD be performed/administered	<i>Benefit >> Risk</i> <i>Additional studies with focused objectives needed</i> IT IS REASONABLE to perform procedure/administer treatment	<i>Benefit ≥ Risk</i> <i>Additional studies with focused objectives needed</i> <i>registry data would be helpful</i> Procedure/Treatment MAY BE CONSIDERED
Estimate of Certainty (Precision) of Treatment Effect	LEVEL A	<ul style="list-style-type: none"> • Recommendation that procedure or treatment is useful/effective • Sufficient evidence from multiple randomized trials or meta-analyses 	<ul style="list-style-type: none"> • Recommendation in favor of treatment of procedure being useful/effective • Some conflicting evidence from multiple randomized trials or meta-analyses 	<ul style="list-style-type: none"> • Recommendation in favor of treatment of procedure being useful/effective • Greater evidence from multiple randomized trials or meta-analyses
	LEVEL B	<ul style="list-style-type: none"> • Recommendation that procedure or treatment is useful/effective • Evidence from single 	<ul style="list-style-type: none"> • Recommendation in favor of treatment of procedure being useful/effective • Some conflicting 	<ul style="list-style-type: none"> • Recommendation in favor of treatment of procedure being useful/effective • Greater evidence from multiple randomized trials or meta-analyses

A		SIZE OF TREATMENT EFFECT		
	Data derived from a single randomized clinical trial or nonrandomized studies	randomized trial or nonrandomized studies	evidence from single randomized trial or nonrandomized studies	randomized trial or nonrandomized studies
	LEVEL C Very limited population evaluated* Only consensus opinion of experts, case studies or standard of care.	<ul style="list-style-type: none"> Recommendation that procedure or treatment is useful/effective Only expert opinion, case studies, or standard-of-care 	<ul style="list-style-type: none"> Recommendation in favor of treatment of procedure being useful/effective Only diverging expert opinion, case studies, or standard-of-care 	<ul style="list-style-type: none"> Recommendation that procedure or treatment is less useful/effective Only diverging expert opinion, case studies, or standard-of-care

*Data available from clinical trials or registries about the usefulness/efficacy in different subpopulations, such as gender, age, history of diabetes, history of prior myocardial infarction, history of heart failure, and prior aspirin use. A recommendation with Level of Evidence B or C does not imply that the recommendation is weak. Many important clinical questions addressed in the guidelines do not lend themselves to clinical trials. Even though randomized trials are not available, there may be a very clear clinical consensus that a particular test or therapy is useful or effective.

Note: In 2003, the American College of Cardiology/American Heart Association (ACC/AHA) Task Force on Practice Guidelines developed a list of suggested phrases to use when writing recommendations. All guideline recommendations have been written in full sentences that express a complete thought, such that a recommendation, even if separated and presented apart from the rest of the document (including headings above sets of recommendations), would still convey the full intent of the recommendation. It is hoped that this will increase readers' comprehension of the guidelines and will allow queries at the individual recommendation level. (See Table 1 in the original guideline document for a list of suggested phrases for writing recommendations.)

CLINICAL ALGORITHM(S)

None provided

EVIDENCE SUPPORTING THE RECOMMENDATIONS

TYPE OF EVIDENCE SUPPORTING THE RECOMMENDATIONS

The type of supporting evidence is identified and graded for each recommendation (see "Major Recommendations").

BENEFITS/HARMS OF IMPLEMENTING THE GUIDELINE RECOMMENDATIONS

POTENTIAL BENEFITS

Appropriate management of patients with tetralogy of Fallot

POTENTIAL HARMS

Despite overall excellent hemodynamic outcomes after surgery for tetralogy of Fallot, there remains a concerning incidence of unexpected sudden death during long-term follow-up (see Table 15 in the original guideline document). Ventricular tachycardia appears to be the mechanism responsible for most of these events, although rapidly conducted intra-atrial re-entrant tachycardia (atrial flutter) or atrioventricular block may be responsible in some cases. The incidence of sudden death for the adult tetralogy population can be estimated from several large series to be on the order of 2.5% per decade of follow-up.

QUALIFYING STATEMENTS

QUALIFYING STATEMENTS

- These practice guidelines are intended to assist healthcare providers in clinical decision making by describing a range of generally acceptable approaches for diagnosis, management, and prevention of specific diseases or conditions. Clinicians should consider the quality and availability of expertise in the area where care is provided. These guidelines attempt to define practices that meet the needs of most patients in most circumstances. The recommendations reflect a consensus of expert opinion after a thorough review of the available current scientific evidence and are intended to improve patient care.
- Patient adherence to prescribed and agreed upon medical regimens and lifestyles is an important aspect of treatment. Prescribed courses of treatment in accordance with these recommendations are only effective if they are followed. Because lack of patient understanding and adherence may adversely affect outcomes, physicians and other healthcare providers should make every effort to engage the patient's active participation in prescribed medical regimens and lifestyles.
- If these guidelines are used as the basis for regulatory or payer decisions, the goal is quality of care and serving the patient's best interest. The ultimate judgment regarding care of a particular patient must be made by the healthcare provider and the patient in light of all of the circumstances presented by that patient. There are circumstances in which deviations from these guidelines are appropriate.

IMPLEMENTATION OF THE GUIDELINE

DESCRIPTION OF IMPLEMENTATION STRATEGY

An implementation strategy was not provided.

IMPLEMENTATION TOOLS

Slide Presentation

For information about [availability](#), see the "Availability of Companion Documents" and "Patient Resources" fields below.

INSTITUTE OF MEDICINE (IOM) NATIONAL HEALTHCARE QUALITY REPORT CATEGORIES

IOM CARE NEED

Getting Better
Living with Illness

IOM DOMAIN

Effectiveness
Patient-centeredness

IDENTIFYING INFORMATION AND AVAILABILITY

BIBLIOGRAPHIC SOURCE(S)

Warnes CA, Williams RG, Bashore TM, Child JS, Connolly HM, Dearani JA, del Nido P, Fasules JW, Graham TP, Hijazi ZM, Hunt SA, King ME, Landzberg MJ, Miner PD, Radford MJ, Walsh EP, Webb GD, Smith SC Jr, Jacobs AK, Adams CD, Anderson JL, Antman EM, Buller CD, Creager MA, Ettinger SM, Halperin JL, Hunt SA, Krumholz HM, Kushner FG, Lytle BW, Nishimura RA, Page RL, Riegel B, Tarkington LG, Yancy CW. Tetralogy of Fallot. In: ACC/AHA 2008 guidelines for the management of adults with congenital heart disease. J Am Coll Cardiol 2008;52(23):e215-21.

ADAPTATION

Not applicable: The guideline was not adapted from another source.

DATE RELEASED

2008

GUIDELINE DEVELOPER(S)

American College of Cardiology Foundation - Medical Specialty Society
American Heart Association - Professional Association

SOURCE(S) OF FUNDING

The American College of Cardiology Foundation and the American Heart Association. No outside funding accepted.

GUIDELINE COMMITTEE

American College of Cardiology/American Heart Association Task Force on Practice Guidelines

Writing Committee to Develop Guidelines on the Management of Adults With Congenital Heart Disease

COMPOSITION OF GROUP THAT AUTHORED THE GUIDELINE

Task Force Members: Sidney C. Smith, Jr, MD, FACC, FAHA, *Chair*; Alice K. Jacobs, MD, FACC, FAHA, *Vice-Chair*; Cynthia D. Adams, RSN, PhD, FAHA#; Jeffrey L. Anderson, MD, FACC, FAHA#; Elliott M. Antman, MD, FACC, FAHA**; Christopher E. Buller, MD, FACC; Mark A. Creager, MD, FACC, FAHA; Steven M. Ettinger, MD, FACC; Jonathan L. Halperin, MD, FACC, FAHA#; Sharon A. Hunt, MD, FACC, FAHA#; Harlan M. Krumholz, MD, FACC, FAHA; Frederick G. Kushner, MD, FACC, FAHA; Bruce W. Lytle, MD, FACC, FAHA#; Rick A. Nishimura, MD, FACC, FAHA; Richard L. Page, MD, FACC, FAHA; Barbara Riegel, DNSc, RN, FAHA#; Lynn G. Tarkington, RN; Clyde W. Yancy, MD, FACC, FAHA

Committee Members: Carole A. Warnes, MD, FRCP, FACC, FAHA, *Co-Chair*; Roberta G. Williams, MD, MACC, FAHA, *Co-Chair*; Thomas M. Bashore, MD, FACC; John S. Child, MD, FACC, FAHA; Heidi M. Connolly, MD, FACC; Joseph A. Dearani, MD, FACC*; Pedro del Nido, MD; James W. Fasules, MD, FACC; Thomas P. Graham, Jr, MD, FACC†; Ziyad M. Hijazi, MBBS, MPH, FACC, FSCAI‡; Sharon A. Hunt, MD, FACC, FAHA; Mary Etta King, MD, FACC, FASE§; Michael J. Landzberg, MD, FACC; Pamela D. Miner, RN, MN, NP; Martha J. Radford, MD, FACC; Edward P. Walsh, MD, FACC||; Gary D. Webb, MD, FACC¶

*Society of Thoracic Surgeons representative.

†International Society for Adult Congenital Heart Disease representative.

‡Society for Cardiovascular Angiography and Interventions representative.

§American Society of Echocardiography representative.

||Heart Rhythm Society representative.

¶Canadian Cardiovascular Society representative.

#Former Task Force member during this writing effort.

**Immediate past chair.

FINANCIAL DISCLOSURES/CONFLICTS OF INTEREST

The American College of Cardiology/American Heart Association (ACC/AHA) Task Force on Practice Guidelines makes every effort to avoid actual, potential, or perceived conflicts of interest that might arise as a result of industry relationships or personal interests among the writing committee. Specifically, all members of the writing committee, as well as peer reviewers of the document, are asked to disclose all such relationships that might be perceived as real or potential conflicts

of interest. Writing committee members are also strongly encouraged to declare previous relationships with industry that might be perceived as relevant to guideline development. If a writing committee member develops a new relationship with industry during their tenure, they are required to notify guideline staff in writing. These statements are reviewed by the parent task force, reported orally to all members at each meeting of the writing committee, and updated and reviewed by the writing committee as changes occur.

Author Relationships With Industry and Other Entities–ACC/AHA 2008 Guidelines for the Management of Adults With Congenital Heart Disease

Committee Member	Research Grant	Speakers' Bureau	Stock Ownership	Board of Directors	Consultant/Advisory Member
Dr. Carole A. Warnes (Co-Chair)	None	None	None	None	None
Dr. Roberta G. Williams (Co-Chair)	None	None	None	None	None
Dr. Thomas M. Bashore	None	None	None	None	None
Dr. John S. Child	None	None	None	None	None
Dr. Heidi M. Connolly	None	None	None	None	None
Dr. Joseph A. Dearani	None	None	None	None	None
Dr. Pedro del Nido	None	None	None	None	None
Dr. James W. Fasules	None	None	None	None	None
Dr. Thomas P. Graham, Jr	None	None	None	None	None
Dr. Ziyad M. Hijazi	None	None	None	None	<ul style="list-style-type: none"> • AGA Medical
Dr. Sharon A. Hunt	None	None	None	None	None
Dr. Mary Etta King	None	None	None	None	None

Committee Member	Research Grant	Speakers' Bureau	Stock Ownership	Board of Directors	Consultant/Advisory Member
Dr. Michael J. Landzberg	<ul style="list-style-type: none"> • Actelion • AGA • Medical • Myogen • NMT Medical • Pfizer 	None	None	None	None
Dr. Pamela D. Miner	None	None	None	None	None
Dr. Martha J. Radford	None	None	None	None	None
Dr. Edward P. Walsh	None	None	None	None	None
Dr. Gary D. Webb	None	None	None	None	None

This table represents the relevant relationships of committee members with industry and other entities that were reported orally at the initial writing committee meeting and updated in conjunction with all meetings and conference calls of the writing committee during the document development process. It does not necessarily reflect relationships with industry at the time of publication. A person is deemed to have a significant interest in a business if the interest represents ownership of 5% or more of the voting stock or share of the business entity, or ownership of \$10,000 or more of the fair market value of the business entity; or if funds received by the person from the business entity exceed 5% of the person's gross income for the previous year. A relationship is considered to be modest if it is less than significant under the preceding definition. Relationships in this table are modest unless otherwise noted.

See Appendix 2 in the original guideline document for peer reviewer relationships with industry.

ENDORSER(S)

American Society of Echocardiography - Professional Association
Heart Rhythm Society - Professional Association
International Society for Adult Congenital Heart Disease - Disease Specific Society
Society for Cardiovascular Angiography and Interventions - Medical Specialty Society
Society of Thoracic Surgeons - Medical Specialty Society

GUIDELINE STATUS

This is the current release of the guideline.

The guidelines will be reviewed annually by the American College of Cardiology/American Heart Association (ACC/AHA) Task Force on Practice Guidelines and considered current unless they are updated, revised, or withdrawn from distribution.

GUIDELINE AVAILABILITY

Electronic copies: Available in Portable Document Format (PDF) from the [American College of Cardiology \(ACC\) Web site](#); electronic copies are also available in PDF from the [American Heart Association \(AHA\) Web site](#).

Print copies: Available from the American College of Cardiology, Resource Center, 9111 Old Georgetown Rd, Bethesda, MD 20814-1699; (800) 253-4636 (US only).

AVAILABILITY OF COMPANION DOCUMENTS

The following are available:

- ACC/AHA 2008 guidelines for the management of adults with congenital heart disease: executive summary. A report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (Writing Committee to Develop Guidelines for the Management of Adults With Congenital Heart Disease). J Am Coll Cardiol, 2008; 52:1890-1947. Electronic copies: Available from the [American College of Cardiology \(ACC\) Web site](#). Also available in Portable Document Format (PDF) from the [American Heart Association \(AHA\) Web site](#).
- ACC/AHA 2008 guidelines for the management of adults with congenital heart disease. Slide set. 2008. 88 p. Electronic copies: Available from the [American College of Cardiology \(ACC\) Web site](#).
- Methodology manual for ACC/AHA Guideline Writing Committees. Methodologies and policies from the ACC/AHA Task Force on Practice Guidelines. 2006 Jun. 61 p. Electronic copies: Available in PDF from the [American College of Cardiology \(ACC\) Web site](#).

Print copies: Available from the American College of Cardiology, 9111 Old Georgetown Road, Bethesda, Maryland 20814-1699.

PATIENT RESOURCES

None available

NGC STATUS

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